

Rehabilitation Of A Patient With Ectodermal Dysplasia With An Overdenture- A Case Report

Abstract

Individuals affected by ectodermal dysplasia syndromes have abnormalities of the glands, tooth buds, hair follicles, and nail development. In addition to these, ectodermal dysplasia causes anodontia and hypodontia intraorally. Partial or total anodontia results in some loss of function, such as chewing, and affects aesthetics. Prosthodontic rehabilitation can be accomplished with fixed, overdenture, complete, or implant-retained prostheses. For rehabilitation, it is crucial to know the age, number and condition of present teeth, and the state of growth of the patient. A 13-year-old male patient who visited our clinic was treated by a multi-disciplinary team of surgeons, orthodontists, and prosthodontists. An overdenture was planned, and an implant-supported prosthesis was considered for when the patient had finished growing. A tooth supported overdenture was planned for prosthetic rehabilitation after considering his growth and the number and condition of his present teeth.

Key Words

Ectodermal dysplasia, anodontia, overdenture

Introduction

Developmental disturbances of dentition encompass a broad range of disorders and affect its size, shape, structure, number, and growth. Pediatric dental patients with any of these developmental disturbances present with challenging esthetic, functional, and psychological needs.¹ Ectodermal dysplasia (ED) has been well documented in the dental literature,¹⁻³ which shows an incidence of about 7 per 10,000 live births.^{4,5} The condition is marked by a developmental deficiency of hair, sweat glands, nails, teeth, and other ectodermal structures.¹⁻⁴ Hypodontia is the second most common complication and affects approximately 80% of patients.⁶⁻⁸ As a result of hypodontia, or anodontia, the alveolar bone is hypoplastic and the alveolus does not develop; however, normal growth occurs in the jaws and face.^{3,7} All of the above contribute to reduced occlusal vertical dimension (OVD).^{2,8} Therefore, removable prosthodontics is required for the growing child and has been reported to be the most common form of prosthodontic intervention.³ This is generally comprised of conventional complete dentures, complete overdentures, or partial dentures.^{1-3,8} In these cases, the advantages of existing teeth with regard to retention, stability, function, and the phonetics of the denture should be considered. In addition, existing teeth help protect the proprioception mechanism and prevent the formation of residual alveolar

ridges. Prosthetic rehabilitation of patients with ectodermal dysplasia is a routine process. This clinical report describes a combined surgical, orthodontic, and prosthodontic approach to the treatment of a patient with anhydrotic ectodermal dysplasia

Case Report

A 13-year-old boy with partial anodontia visited our clinic because he had difficulty chewing. The patient had been diagnosed with anhydrotic ectodermal dysplasia when he was 3-years-old.

The patient had the typical characteristics of anhydrotic ectodermal dysplasia, such as protuberant lips, a saddle nose, fine sparse hair, and scant eyelashes and eyebrows. (Fig 1)



Fig 1 - Frontal View

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He had hyperkeratosis on his palms and soles. His parents lacked these characteristics, while his teenage sister had some of the characteristics, such as a saddle nose, sparse hair, prominent supraorbital ridges, and conical, hypoplastic front teeth. Intra-oral examination revealed a dry oral mucosa, protuberant lips, a thin vermilion line, and undeveloped alveolar ridges. There were two conical, hypoplastic anterior teeth in the right maxilla. The mandible was also devoid of ant teeth except for a first molar on the right side.

Prosthodontic Treatment

A diagnostic cast was prepared and the denture was planned. We decided to apply a complete denture for the mandible and an overdenture for the maxilla. In the evaluation, the coronal structures of the existing teeth, axial wall inclination, and relative positions were considered to determine the path of insertion, retention, and stabilisation of the overdenture, to minimize plaque accumulation on the hypoplastic teeth, and to optimise the crown/root ratio of the fabricated metal copings. While preparing the teeth for the metal copings, the slopes of the axial walls were made nearly parallel. (Fig 2) An impression was taken with an individual tray using medium-viscosity silicone impression material (Xantopren® M, Bayer Dental, Germany). The copings were cast with a Cr-Ni-based metal alloy (Wiroloy®, BEGO,

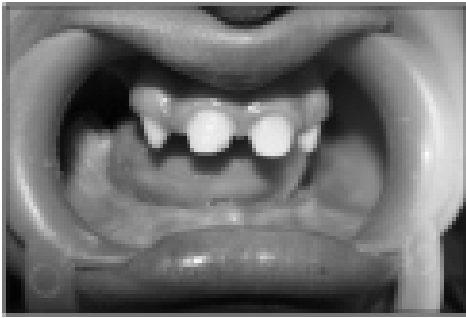


Fig 2 - Tooth Preparation

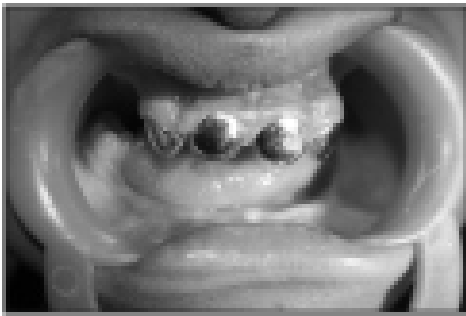


Fig 3 - Metal Copings



Fig 4 - Final Denture

occurred.

Discussion

Ectodermal dysplasia syndrome was first described in the medical literature by Thurnam, who reported two typical patients in 1848.¹ Individuals affected by ectodermal dysplasia syndromes have abnormalities of the glands, tooth buds, hair follicles, and nail development. Some ectodermal dysplasia syndromes are mild, while others are devastating. Other symptoms include deficient tears and saliva, poorly functioning mucous membranes, frequent respiratory infections, hearing or vision deficits, missing fingers or toes, cleft lip or palate, problems with the immune system, sensitivity to light, lack of breast development, and other abnormalities of the ectoderm. Lifespan can be affected in some rare ectodermal dysplasia syndromes.^{1,2} However, there are very few documented examples of a person affected by ectodermal dysplasia syndrome dying because of an inability to perspire. Anhydrotic ectodermal dysplasia is a rare X-linked condition. Affected males lack teeth, have hypotrichosis, and no sweat glands. Patches with and without sweat glands are seen in heterozygous females. Anhydrotic ectodermal dysplasia is a triad of hypodontia or anodontia, hypotrichosis,⁴ and hypohidrosis, and is associated with other problems that result from the defective development of structures of ectodermal origin.⁵ This is one of 132 known clinical ectodermal dysplasia syndromes.²

Anhydrotic ectodermal dysplasia occurs in all races, with an incidence of 1 to 7 per 100,000 live births.³ Patients with anhydrotic ectodermal dysplasia generally have prominent supraorbital ridges, frontal bossing, and a saddle nose.¹⁵ The maxillae may be underdeveloped and the lips thick and prominent. The nose may appear pinched, and the alae nasi hypoplastic. The patient may resemble an edentulous old person. Some patients do not produce tears. The nails are usually normal. The skin of an infant may appear hypopigmented. Maculopapular eruptions may occur during infancy. Asthma and eczema are occasionally reported in these patients.¹⁸ The characteristic dental defect in this syndrome is peg-shaped or conical front teeth, which cannot be distinguished from incisors. Both the deciduous and permanent teeth are affected. Anodontia may occur, but hypodontia with misshapen teeth is usual, and these teeth may be hypoplastic.^{1,2,3} Early and extensive dental treatment is needed

throughout childhood because of the absence of most of the deciduous and permanent dentition.

A multidisciplinary team approach to treatment is recommended in these cases.¹⁰ Osseo-integrated implants are an alternative treatment in older people with anhydrotic ectodermal dysplasia.^{13,14} Considering the age and potential growth of our patient, it was deemed better to postpone osseointegrated implants. As the physical development of our 10-year-old male patient was incomplete, transitional over denture was planned, which allowed us to make adjustments. Osseointegrated implants and permanent prosthodontic treatment are planned for when the patient is fully grown.

Transitional overdentures are advantageous from the perspective of psychological development, and with patients feeling more secure with the aesthetic appearance of an overdenture.

Conclusion

The use of partial acrylic prostheses is an interesting and practical alternative that provides a relatively quick, easy, acceptable and economical solution to the functional and esthetic oral rehabilitation in patients with pronounced edentulism. This solution improves the patient's quality of life and optimizes social integration; furthermore, it permits stimulation of the alveolar ridges for later treatment with an implant supported prosthesis as a more stable and esthetic solution for patients with multiple dental agenesis.

References

1. Nyhan WL. Diagnostic recognition of genetic disease. 1st ed. pp 680-685. Philadelphia: Lea & Febiger, 1987.
2. Jones KL. Smith's recognizable patterns of human malformation. 4th ed. pp 254-255. Philadelphia: WB Saunders, 1988.
3. Casamassimo PS, McTigue DJ, Fields HW, Nowak A. Pediatric dentistry (Infancy through adolescence). 3rd ed. pp 412-413. Philadelphia: WB Saunders, 1999.
4. Blattner RJ. Hereditary ectodermal dysplasia. J Pediatr 1968; 73: 444-447.
5. Berg D, Weingold DH, Abson KG, Olsen EA. Sweating in ectodermal dysplasia syndromes: A review. Arch Dermatol 1990; 126: 1075-1079.
6. Feire-Maia N, Pinheiro M. Ectodermal dysplasia-some recollections and a classification. Birth Defects 1988; 24: 3-

Germany).

The copings were cemented to the teeth using polycarboxylate cement (**Fig 3**) (Harvard CC carboxylate cement, Harvard Dental-GmbH, Berlin, Germany). As the patient had a dry mucosa, the impression was taken using rapid-setting silicone impression material with high elasticity.^{14, 15} Bilateral balanced occlusion was developed using anatomic (33 degree cusp angle) acrylic teeth. The maxillary overdenture and complete mandibular denture were prepared by conventional methods using heat-polymerizing acrylic resin. (**Fig 4**) The patient was seen 48 hours later for adjustment, then at one and two weeks and 1, 3, and 6 months. The patient was followed up annually. No major complications

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7. Buyse ML. Birth defects encyclopedia.. pp 597-8. Chicago-St Louis: Mosby, 1990.
8. McDonald RE, Avery DR. Dentistry for the child and adolescent..7th ed. pp 134-7. St. Louis: Mosby Inc,1999.
9. Farrington FH. The team approach to management of ectodermal dysplasia.Birth Defects 1988; 24: 237-242.
10. Guckes AD, Brahim JS, McCarthy GR, Rudy SF, Cooper LF. Using endosseous dental implants for patients with ectodermal dysplasia. J Am Dent Assoc1991; 122: 59-62.
11. Pigno MA, Blackman RB, Cronin RJ Jr, Cavazos E. Prosthodontic management of ectodermal dysplasia: a review of the literature. J Prosthet Dent 1996;76: 541-545.
12. Kearns G, Sharma A, Perrott D, Schmidt B, Kaban L, Vargervik K. Placement of endosseous implants in children and adolescents with hereditary ectodermal dysplasia. Oral Surg Oral Med Oral Pathol Oral Radio Endod 1999;88: 5-10.
13. Anusavice KJ. Phillips' science of dental materials. 10th ed. pp 162. Philadelphia: WB Saunders, 1996.
14. O'Brien WJ. Dental materials and their selection. 2nd ed. pp 134. Carol Stream: Quintessence, 1997.
15. Clarke A. Hypohydrotic ectodermal dysplasia. J Med Genet 1987; 24: 959-956.

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